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RELIABILITY OF THE LAPAROSCOPIC IDENTIFICATION OF THE LEVEL OF TRANSITIONAL ZONE IN HIRSCHSPRUNG`S DISEASE

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ABSTRACT

Background/Aim: It is crucial to exactly identify the level of transition to the normal ganglionic colon in instances of Hirschprung's disease (HD). The correlation between the laparoscopic leveling of the transitional zone (TZ) and frozen section histopathologically based level of aganglionosis has not been well studied. The aim of this study is to assess whether the laparoscopic leveling of TZ could be a reliable method for the diagnosis of the actual level ofaganglionosis of the HD. **Patients and Methods:** The study included 150 patients' diagnosedas HD by rectal biopsy. All patients were subject to laparoscopy for identification of TZ. Three laparoscopic seromuscularbiopsies was taken for intra-operative frozen section [IOFS] from thelaparoscopically identified TZ, 2-cm distal and 10-cm proximal to TZ. In cases of non-identification of any TZ, laparoscopic seromuscular biopsies were taken from therectosigmoid, sigmoid, descending colon, splenic flexure and the transverse colon for IOFS. Then, laparoscopic assisted endo-rectal pull through was completed according to the results of IOFS. The results of IOFS were compared with the results of permanent paraffin section **Results:** The study included 98 males and 52 females with HD. Their mean age was 2.4±12 years (age range = 5 months to 6 years). All operations were completed laparoscopically without conversion. The mean operative time was 54.4±12 minutes (range =45 to 70 minutes). Laparoscopic identification of the TZ coincided with the suspected TZ in the images of preoperative barium enema in 125 patients. While in 25 patients without a detectable TZ in the preoperative barium enema, TZ was identified laparoscopically. Laparoscopic identification of the level of the TZ zone coincided with thehistopathological results of IOFS [IOFS] and permanent paraffin section in all patients. No intraoperative complications were reported. **Conclusion:** Laparoscopic identification of TZ in patients with HD is a feasible and a safe procedure. It is a reliable method for determining the actual level of aganglionosis in HD.

KEYWORDS: Laparoscopy, Frozen section, Biopsy, Hirschsprung`s disease. Leveling colostomy. Transitional Zone.

INTRODUCTION

Hirschsprung's disease (HD) is a relatively common disease, occurring in approximately 1 in 5,000 live births. The majority of cases present during the first few months of life with delayed passage of meconium, distal intestinal obstruction, chronic constipation, abdominal distension, or unexplainable enterocolitis.[1] There is a trend toward primary resection without prior colostomy in the surgical treatment of HD that renders evaluation of the initial pathologic specimen of utmost importance. The anticipated level of aganglionosis can influence the surgical approach to HD.[2] The definitive treatment of HD depends mainly on leveling biopsies. Open leveling biopsies carry many complications as similar to that of the definitive repair. Interpretation of the intraoperative frozen section [IOFS] is not available in every center and although the rate of its error could be very low, but it carries a significant impact on the patients, when it occurs.[1,3] Although the detection of a TZ by the laparoscopy may aid in the diagnosis of HD, the correlation between the location of the TZ and the pathologic level of aganglionosis has not been well studied. Can we depend on the laparoscopic appearance of TZ and dilated colonic segment for leveling of HD to avoid the patient from the complications ofmulti-staged operations?.We, therefore, sought to determine the accuracy of the laparoscopic identification of the TZ in comparison with the histopathological results of IOFS and the histopathologyParaphene examination of the resected specimen in detecting the level of aganglionosis.

PATIENTS AND METHODS

The study was conducted at Al-Azhar University Hospitals and some private Hospitals during the period from January 2012 to December 2014. It included 150 patients with clinically suspected HD, barium enema suspicious of HD and aganglionicrectal biopsy. All patients were subject to thorough clinical examination, laboratory investigations, barium enema, and rectal biopsy.

All patients were subject to laparoscopic evaluation of the rectum, sigmoid colon, descending colon, and transverse colon for identification of the TZ.

Where is mention about the Medical Ethics Committee and the medical consent from the patients?

Main outcome measurements included: Safety of the procedure, operative time, hospital stay and laparoscopic appearance of TZ and its correlation with the results of frozen section and histopathology of the resected colonic specimen.The study was approved by the Ethical Committee of our hospital. All parents signed a written detailed informed consent.

All patients were subject to colonic preparation by twice daily warm saline enema (20 ml/kg) for one week preoperatively. They received one doseof IV 3rd generation Cephalosporin, 50 mg/kg body weight, in the morning of surgery.

Operative procedure: After induction of general endotracheal tube anesthesia, the patient was placed supine in the Trendelenburg`s position.A 5-mm port was inserted at the umbilicus by open Hasson`s technique. Through this port, pneumoperitoneum was created to 10-12 mmHg then, a 5-mm scope 30 degree was used. Two 3-mm accessory trocars were inserted under direct vision at the lateral edge of the right and leftrectus abdominis muscle at the level of umbilicus.

Traction of the colon by 2 stay sutures inserted percutaneously through the anterior abdominal wall 8-10 cm. apart. A curved needle with prolene suture No. 0. mounting on a heavy needle holder is inserted from outside penetrating the abdominal wall, piercing the seromuscular layer of the colon and passes again through the abdominal wall to outside.Both sutures are used in controlling the retraction of the colon by pulling and relaxing the thread according to the need.

The first step of the laparoscopic procedure was the complete visualization of the abdomen and pelvis. Then, the rectum, sigmoid colon,then the descending and the transverse colon for identification of any TZ. When a TZ has been identified, seromuscular stitch was applied as a landmark above its proximal limit (nearly about 4-cm proximal to the upper level of TZ).

In cases of inability to identify a clear visualization of the TZ, laparoscopic seromuscular biopsies were taken from rectosigmoid, sigmoid, descending colon, splenic flexure, transverse colon and the biopsy sites were closed with intracorporeal interrupted stitches.The definitive surgical management of these cases was tailored according to the results of the histopathology of the biopsies.The laparoscopic technique was described earlier.[4,5] In short, the extension of the aganglionosis was diagnosed by laparoscopic identification of TZ.

In patients with classic forms of TZ, the peritoneal reflection was incised circumferentially, and mobilization started at the rectosigmoid down to the pelvic floor. Then, dissection of the bowel in the proximal direction was carried and the mesenteric vessels were clipped and divided leaving the marginal vessels intact until the colon pedicle was long enough to reach deep into the pelvis without tension. The anterior wall of the rectum was not dissected, and the posterior plane was dissected 2–3 cm distal into the pelvis, hugging the rectal wall.

Transanal dissection was started by placing six to eight traction sutures or Lone Star retractor to retract the anus. A circumferential incision was made in the rectal mucosa at 0.5–1 cm above the dentate. Rectal mucosal dissection was performed transanally by mono-polar diathermy. A submucosal plane was developed and extended superiorly for more than 5 cm until the peritoneal reflection, in an effort to prevent injury to the pelvic tissues (Fig. 3). To avoid injury to the ureter, vas deferens, and/or perirectal structures, endorectal dissection was continued upward in a circumferential manner until the level of the laparoscopic dissection. Once the colon was pulled through the anus up to the site of the landmark stitch, the laparoscope was again used to ensure that the bowel was not twisted. The muscular cuff was then shortened to 2–3 cm. The aganglionic and dilated segments were resected and a coloanal anastomosis was performed with interrupted fine absorbable sutures. The laparoscopic leveling of the TZ was compared with the histopathological results of the resected colonic specimen.

RESULTS

A total of 150 patients, were included in this study. They were 98 males and 52 females. Demographic data were obtained from hospital records. Their mean age was 2.4±12 years old (range= 5 months to 6 years). Definite laparoscopic findings were present in all cases in the form of; clear peritoneal reaction, redundancy of the rectosigmoid colon, lymph node enlargement in the mesentery of the rectosigmoid. No intraoperative complications were reported. Early postoperative complications, mainly perianal excoriation, occurred in 2 patients. Demographic data and the laparoscopic finding are shown in [Table 1]. The mean operative time was 54.4±12 minutes (range= 45 to 90 minutes). Mean postoperative hospitalization was 4 days (range =3 to 7days). One case developed postoperative enterocolitis and was managed by colon irrigations, IV fluids and antibiotics. One case developed anastomotic leak and was treated by colostomy. There was no recurrent constipation in both groups.

Laparoscopic TZ was identified at the rectosigmoid junction and sigmoid colon in 121 patients, lower third of the descending colon in 17 and at middle third of the descending colon in 7 patients [Fig. 1, 2]. While in 5 patients, laparoscopic TZ was not apparent and the

results of the histopathological studies of the biopsies in these patients were ganglionic.

Laparoscopic identification of the TZ level coincided with the images of the preoperative barium enema in 125 cases. While in 25 patients without a detectable TZ in the preoperative barium enema, TZ was identified laparoscopically.

The laparoscopic view of the TZ coincided with the results histopathology of frozen section in all cases. All the patients achieved full recovery of their symptoms at the late post-operative follow up after one year. Three cases developed soiling which improved spontaneously within 3 monthsThe cosmetic results were excellent.[Fig.4].

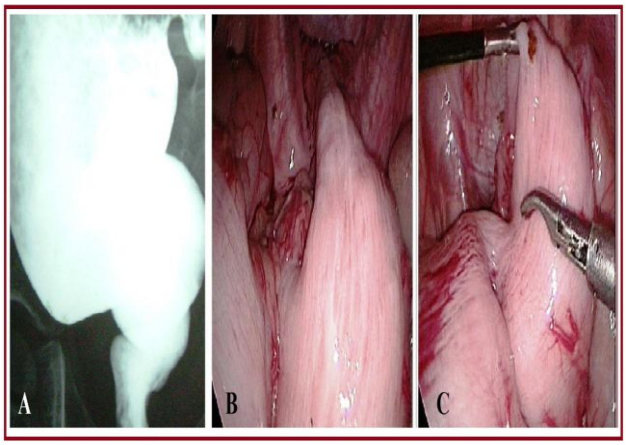

Figure 1: A) Barium enema showing typical case of rectosigmoidHirschsprung`s disease. B –C) Laparoscopic View of TZ at the rectosigmoid junction.

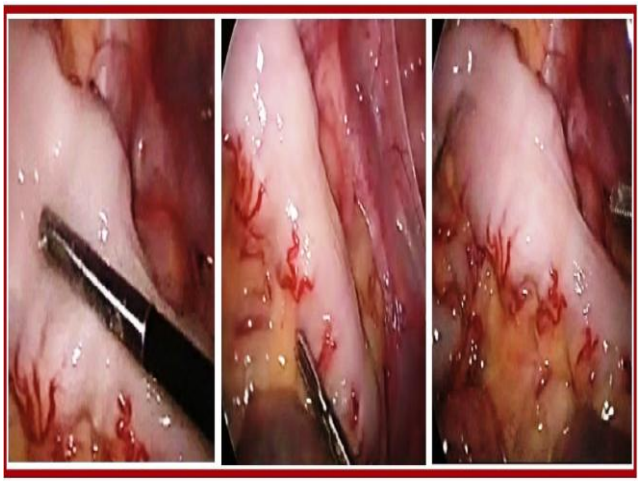

Figure 2: Laparoscopic View of TZ at the descending colon.

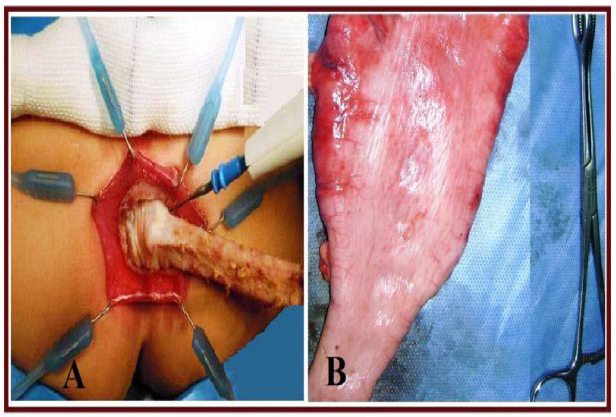

Figure 3: A) Showing transanal Endorectal pull-through with Lone Star retractor in place. B) Showing specimen removed for a case of rectosigmoidHirschsprung`s disease.

Figure 4: Showing excellent cosmetic Results.

DISCUSSION

Major advances have occurred in the management of HD since Swenson described his definitive operation in 1948. In spite of these advances, still there is a great controversy in the management of HD. Traditionally; surgical correction of HD has involved a 2- or 3-stage approach in which the level of aganglionosis is confirmed by open full-thickness biopsy at the time of initial leveling colostomy followed later on by a definitive pull-through procedure. More recently, single-stage repair has been shown to be safe and effective approach, with the benefit of eliminating the need for colostomy and reducing the number of hospitalizations.[6,7,9,5]

The development of laparoscopic-assisted endorectal pull-through for Hirschsprung's disease currently offers a minimally invasive alternative to open abdominal surgery. It offers the additional advantages of reduced morbidity, improved cosmoses, decreased postoperative pain, and shorter postoperative hospital stay in comparison with open procedures.[8,9,10,11]

The laparoscopic approach has several important advantages over the traditional abdominal approach. It providesclear delineation of pelvic structures, faster postoperative recovery, and better cosmetic results.[9,10]

Laparoscopy can also identify the level of normal innervation, as suggested by the transitional zone. It can avoid twisting and tension of the pull-through segment and overcome the difficulty in dividing mesenteric vessels compared with the pure transanal approach.[5,12] However, it is crucial to accurately identify the exact level of transition to the normal colon in instances of HD. The correlation between the location of the radiological transitional zone [RTZ] and the pathologic level of aganglionosis has not been well studied. With the advent of a single-stage approach for the surgical management of HD, the barium enema has been increasingly relied on to predict the level of aganglionosis and, in some cases, determine the approach of surgical procedure.[13] Many radiographic features in the barium enema suggest the diagnosis of HD including; the presence of TZ, abnormal narrowing of the distal aganglionic segment and delayed evacuation of barium after 24hours. Barium enema, in the majority of cases may aid in the diagnosis of HD. The presence of RTZ in barium enema remains the most reliable diagnostic tool.[14,15] However, several investigators have reported cases in which the radiologic and pathologic transition zones differ. Some investigators have found that RTZ correlates well with the level of aganglionosis and depend on the picture of barium enema in identification of the level of resection during the definitive surgery, but they have 4 % recurrence rate of constipation.[16] Thus, many surgeons believe that, the TZ should be confirmed either laparoscopically or by an IOFS through a small umbilical incision, should be performed before beginning the rectal dissection.[6] It is incumbent on pathologists, surgeons, and institutions caring for children with this disease to take all steps necessary to minimize the incidence of pathologic error by paying close attention to technical issues during the performance and preparation of the biopsies. Potentially confounding factors that may have contributed to the misdiagnosis on IOFS included immature ganglion cells, technical difficulty and freezing artifact, limited experience of the pathologist, and superficial sectioning in frozen section slides.[1]

In this study we correlated the laparoscopic finding with the clinical examination, barium enema, IOFS and histopathological results. Our results coincided with that of the others.[1,15,17,18] In our study, the laparoscopic view of TZ correlated accurately with barium enema in 83.3 % of cases.[19,20,21] Proctor et al stated that, 90% of patients with RTZ in the rectosigmoid junction correctly predicted the level of aganglionosis in 89% of patients with rectosigmoid disease but only 31% of those with long-segment or total colonic disease.[2] Laparoscopic identification of the view of TZ, correlated accurately with IOFS and the histopathological study of the resected colonic specimens in all cases. The authors have adopted a laparoscopic approach that allows the surgery to be performed in one stage with a marked decrease in the morbidity and the hospital stay.

Although the light microscopy features of ganglion cells IOFS are readily notable in the majority of cases, the use of IOFS is not available in many cases.[3] Shayan et al stated that, the rate of error during IOFS in children with HD can be very low, but when it occurs, it has a significant impact on the patients.[1,3,22] In this study, the laparoscopic view of the TZ was used for the diagnosis of the level of HD after correlation with IOFS and histopathological study of the resected specimen. When the laparoscopic view of TZ is not clear, laparoscopic seromuscular biopsies were taken from rectosigmoid, sigmoid, descending colon, splenic flexure, transverse colon and the defect was closed with intracorporeal interrupted stitches. According to the results of histopathology, the definitive treatment of HD was accomplished by either trans-anal endorectal pull-through or by laparoscopic assisted Duhamel`s procedures By this method, we have greatly excluded a chance of having any residual aganglionic segment. In a series of 80 patients with HD reported by Maia , the concordance rates between IOFS and permanent section was 67% for initial pathologic specimens (ie, rectal biopsies) and overall rate of 89% [3] However, Shayan et al found a higher concordance rate between IOFS and permanent section results reaching to 98.7%. Other surgeons use laparoscopy for the identification of the region of caliber change and division of restrictive mesenteric vascular branches before starting the perineal procedure.[1,23,24]

The surgical management of HD has evolved over the last 2 decades. The traditional approach included a laparotomy with serial biopsies, using IOFS to identify the transition zone and to form a colostomy at the point at which ganglion cells are identified. The IOFS diagnosis was then confirmed on permanent sections and after few months, the definitive "pull-through" procedure was performed. In recent years, increasing numbers of pediatric surgeons have abandoned the routine use of a colostomy and have performed a 1-stage pull-through procedures. Because this requires the use of IOFS to determine the level of TZ, an error in diagnosis can have a significant negative impact on the clinical outcome.[25] One controversy which continues to be pertinent is whether the transanal dissection should be initiated before confirming the location of the pathologic transition zone.

Over the past few years, the popularity of minimal-access surgical techniques has led to a number of modifications to the standard 1-stage operations. Georgeson described a laparoscopic approach that has been adopted by many surgeons and that is associated with less pain and a shorter hospitalization than standard open procedures.[8] Subsequently, a number of authors

described a solely trans-anal approach, which appears to be associated with the same advantages without the need for any intraabdominal dissection.[25,26]

The first series of single-incision laparoscopic endorectal pull- through (SILEP) in July 2010 involved six patients with HD reported by Muensterer and it was found to be safe andfeasible.[27]

Yamataka et al developed a simple technique for laparoscopy assisted suction colonic biopsy (SCBx) during trans-anal pull-through for rectosigmoid HD. It accurately identifies the exact level of normo-ganglionosis by naked eye appearance confirmed by histology. After identification of the region of caliber change by laparoscopy, the suction biopsy device was inserted through the anus ensuring that the tissue sampling mechanism lay anteriorly so the laparoscopic team could check for any perforation at the time of biopsy.[6]

Some surgeons identify the level of normo-ganglionosis by performing a full-thickness biopsy of the rectum or colon when it is prolapsed through the anus after the region of caliber change is identified laparoscopically[4,8,9] However, others prefer to determine the level of normo-ganglionosis histologically before the bridge-burning part of the operation, which is, the anorectal dissection from below. Shams[28] reported an accuracy of the laparoscopic view with the result of biopsy in a study addressing the value of laparoscopic assisted biopsy.

In the laparoscopic colon pull-through, one can remove not only the aganglionic segment and transitional segment, but also the malfunctioned dilated segment. This may further reduce the rate of postoperative enterocolitis, as the bowel segment used in anastomosis has a normal caliber, and the coloanal anastomosis can be easily performed.

Vũused suture string to gain exposure to the anus, until 2 years ago when they turned to use Lone Star retractors, which, had better anal exposure. The soiling and increased frequency of bowel movements in several patients may be due to dissection too close to the dentate line or overstretching of the pulled colon.[29,30] Conclusion: Laparoscopic identification of TZ is feasible, safe, decisive technique for diagnosing and leveling of HD primarily without colostomy. It saves the patient the complications of multiple stages operations, multiple anesthesia. It is a reliable method in all cases if the TZ was apparent during laparoscopy.

REFERENCES

1. Shayan K, Smith C, Langer JC: Reliability of intraoperative frozen sections in the management of Hirschsprung's disease J Pediatr Surg, 2004; 39(9): 1345-8.
2. Proctor ML, Traubici J, Langer JC et al: Correlation between radiographic transition zone and level of aganglionosis in Hirschsprung's disease: Implications for surgical approach. J Pediatr Surg, 2003; 38: 775-8.
3. Maia DM. The reliability of frozen-section diagnosis in the pathologic evaluation of Hirschsprung's disease. Am J SurgPathol, 2000; 24: 1675-7.
4. Georgeson KE, Fuenfer MM, Hardin WD: Primary laparoscopic pull-through for Hirschsprung's disease in infants and children. J PediatrSurg, 1995; 30: 1017-22
5. Tang ST, Wang GB, Cao GQ et al. 10 years of experience with laparoscopic-assisted endorectal Soave pull-hrough procedure for Hirschsprung'sdisease in China.J Laparoendosc AdvSurg Tech A, 2012; 22: 280-4.
6. Yamataka A, Yoshida R, Kobayashi H, et al Laparoscopy-Assisted Suction Colonic Biopsy and Intraoperative Rapid Acetyl cholinesterase Staining During Trans-anal Pull-Through for Hirschsprung's Disease. J Pediatr. Surg, 2002; 37: 1661-3.
7. So HB, Schwartz DL, Becker JM, et al: Endorectal pull-through without preliminary colostomy in neonates with Hirschsprung's disease. J PediatrSurg, 1980; 15: 470-1.
8. Hackam DJ, Superina RA, Pearl RH: Single-stage repair of Hirschsprung's disease: A comparison of 109 patients over 5 years. J PediatrSurg, 1997: 32: 1028-32.
9. Pierro A, Fasoli L, Kiely EM, et al: Staged pull-through for rectosigmoidHirschsprung's disease is not safer than primary pull-through. J PediatrSurg, 1997; 32: 505-9.
10. Craigie RJ, Conway SJ, Cooper L, et al. Primary pull-through for Hirschsprung's disease: Comparison of open and laparoscopic-assisted procedures. J aparoendosc AdvSurg Tech A, 2007; 6: 809–12.
11. Thomson D, Allin B, Long AM et al. Laparoscopic assistance for primary transanalpullthrough Hirschsprung's disease: systematic review andmeta-analysis. BMJ Open, 2015; 245(3): 103.
12. De la Torre-Mondrago`n L, Langer JC. Transanalendorectal pull-through for Hirschsprung disease: Technique, controversies, pearls, pitfalls, and an organized approach to the management of postoperative obstructive symptoms. SeminPediatrSurg, 2010; 19: 96–106.
13. Hoehner JC, Ein SH, Shandling B, et al: Long-term morbidity in total colonic aganglionosis. J PediatrSurg, 1998; 33: 961-6.
14. Berdon WE, Baker DH: The roentgenographic diagnosis of Hirschsprung's disease in infancy. AJR, 1965; 93: 432- 46
15. Rosenfield NS, Ablow RC, Markowitz RI, et al: Hirschsprung disease: Accuracy of the barium enema examination. Radiology, 1984; 150: 393-400.
16. Elhalaby EA, Hashish A, Elbarbary MM, Soliman HA et al: Transanal one-stage endorectal pull-

through for Hirschsprung's disease: a multicenter study. J Pediatr Surg, 2004; 39: 345-51.
17. Kleinhaus S, Boley SJ, Sheran M, et al: Hirschsprung's disease: A survey of the members of the surgical section of the American Academy of Pediatrics. J PediatrSurg, 1979; 14: 588-97.
18. Taxman TL, Yulish BS, Rothstein FC: How useful is the barium enema in the diagnosis of infantile Hirschsprung's disease? AJDC, 1986; 140: 881-4.
19. Quinlivan JA, Petersen RW, Nicklin JL. Accuracy of frozen section for the operative management of endometrial cancer. Br J Obstet Gynecol, 2001; 108: 798-803.
20. Caraci P, Aversa S, Mussa A. Role of fine-needle aspiration biopsy and frozen- section evaluation in the surgical managenent of thyroid nodules. Br J Surg, 2002; 89: 797-801.
21. King SK, Sutcliffe JR, Hutson JM: Laparoscopic seromuscular colonic biopsies: a surgeon's experience. J Pediatr Surg, 2005; 40: 381-4.
22. Law CHL, Walton JM, Fitzgerald PG: Laparoscopic Swenson's pull-through for Hirschsprung's disease. Pediatr Endosurgery & Innovative Techniques, 1999; 3: 181-185.
23. Morikawa Y, Hoshino K, Matsumura K, et al: Extra-anal mucosectomy: Laparoscopic-assisted endorectal pull-through using a prolapsing technique. J Pediatr Surg, 1998; 33: 1679-81.
24. Langer JC, Fitzgerald PG, Winthrop AL. One vs two stage Soave pull-through for Hirschsprung's disease in the first year of life. J PediatrSurg, 1996; 31: 33-7.
25. Langer JC, Minkes RK, Mazziotti MV, et al. Transanal one-stage Soave procedure for infants with Hirschsprung disease. J Pediatr Surg, 1999; 34: 148-152.
26. Langer JC, Seifert M, Minkes RK. One-stage Soave pullthrough for Hirschsprung disease: a comparison of the transanal vs open approaches. J Pediatr Surg, 2000; 35: 820-2.
27. Muensterer OJ, Chong A, Hansen EN, et al. Single-incision laparoscopic endorectal pull-through (SILEP) for Hirschsprung disease. J GastrointestSurg, 2010; 14: 1950-4.
28. Shamseldin A M. Laparoscopic assisted leveling biopsies for Hirschsprung`s disease. Al-Azhar Med J, 2011; 40: 595- 601.
29. Pratap A, Gupta DK, Shakya VC, et al. Analysis of problems, complications, avoidance and management with transanal pull-through for Hirschsprung disease. J PediatrSurg, 2007; 11: 1869–76.
30. Vũ PA, Thien HH, Hiep PN. Transanal one-stage endorectal pull-through for Hirschsprung disease: Experiences with 51 newborn patients. Pediatr SurgInt, 2010; 26: 589–92.